Clinical research

Microsurgical management of non-neurofibromatosis spinal schwannoma

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ABSTRACT

Introduction: The aim of this study is to assess the clinical properties and surgical results of patients diagnosed with spinal schwannomas without neurofibromatosis (NF) properties.

Patients and methods: The data obtained from 35 patients who underwent resection of spinal schwannomas were analyzed. All cases with neurofibromas and those with a known diagnosis of NF Type 1 or 2 were excluded. 35 patients underwent surgery for spinal schwannoma at our institution between January 1997 and 2010. The data were gathered retrospectively from medical records and included clinical presentation, tumor location and post-operative complications. All cases were surgically excised, and they were confirmed to be schwannomas by pathologists with histopathological sections in paraffin stained with hematoxylin–eosin.

Result: We treated 35 (20 males and 15 females) patients with spinal schwannomas. The mean age of the patients was 47.2 (between 13 and 76) years. Of the cases, six schwannomas were located in the cervical spine, four in the thoracic spine, two in cervico-thoracic area, 10 in the thoraco-lumbar area and 13 in the lumbar spine. Two patients had malignant schwannomas that were recurrent. Of the 35 cases, the schwannomas were intradural–extramedullary in 30 cases (86%), intradural–intramedullar in 2 cases (6%), and extradural in 3 cases (9%).

Conclusion: Spinal schwannomas may occur at any level of the spinal axis and are most frequently intradural–extramedullary. The most common clinical presentation is pain. Most of the spinal schwannomas in non-NF patients can be resected completely without or with minor post-operative deficits. This knowledge may help us to create a strategy for total resection of a spinal schwannomas.

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Gestión microquirúrgica de schwannoma medular sin neurofibromatosis

RESUMEN

Introducción: El objetivo de este estudio es evaluar las características clínicas y los resultados quirúrgicos de un grupo de pacientes con diagnóstico de schwannoma medular sin propiedades de neurofibromatosis (NF).

Pacientes y métodos: Se analizaron los datos de 35 pacientes sometidos a resección de schwannomas medulares. Todos los casos con neurofibromas y aquellos con un diagnóstico conocido de NF tipo 1 o 2 fueron excluidos del estudio. Los 35 pacientes fueron sometidos a cirugía por schwanna medular en nuestro centro, entre enero de 1997 y 2010. Los datos obtenidos retrospectivamente de los registros médicos incluyeron la presentación clínica, localización del tumor y complicaciones post-operatorias. Todos los casos fueron extirpados quirúrgicamente, y confirmados como schwannomas mediante especímenes histopatológicos teñidos con hematoxilina-eosina.

Resultado: Se trató a 35 (20 hombres y 15 mujeres) pacientes con schwannomas medulares. La edad media de los pacientes fue de 47,2 años (rango entre 13 y 76). Seis schwannomas se encontraban en la columna cervical, 4 en la columna torácica, 2 en la zona cérvido-torácica, 10 en la zona torácico-lumbar y 13 en la columna lumbar. Dos pacientes sufrieron recurrencias de schwannomas malignos. De los 35 casos, 30 (86%) fueron schwannomas intradurales-extradurales, 2 (6%) fueron intradurales-intradurales, y 3 (9%) fueron extradurales.

Conclusión: Los schwannomas medulares pueden aparecer a cualquier nivel de la columna vertebral aunque los más frecuentes son los de localización intradural-extradural. La presentación clínica más común es el dolor. La mayoría de los schwannomas medulares en pacientes sin NF pueden ser resecados en su totalidad, con o sin secuelas post-operatorias menores. Este conocimiento puede ayudar en la creación de una estrategia para la resección total de schwannomas medulares.

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Introduction

Schwannomas are one of the most commonly found extradural spinal tumors.1-4 Along with spinal neurofibromas, they are classified as spinal nerve sheath tumors (NSTs).4 In some studies, it has been noted that the classification and nomenclature of NSTs have been perplexing.2,5 It has been suggested that this confusion stems from the erroneous interchangeable use of the words neurofibroma and schwannoma; the frequent use of the inaccurate terms “neuroma,” “neurinoma,” and “neurilemoma,” and the grouping of all spinal NSTs in the same series. Nevertheless, due to their different demographic, histological, biological, and clinical characteristics, neurofibromas and schwannomas deserve separate consideration. NSTs are linked with both NF1 and NF2.4 Even so, neurofibromas are dominant in NF1 and schwannomas are more prevalent in NF2.4-6 Neurofibromatosis Type 2 is connected with the more aggressive biological behavior of schwannomas and has different clinical characteristics.1,7 For this reason, in this study, we excluded all patients with a known diagnosis of NF because the lesions in these groups possess different histological and biological features that may obscure the true clinical characteristics of spinal schwannomas. In this article, we present our 14-year experience in treating 35 patients with spinal schwannomas.

Patients and methods

35 patients underwent surgery for spinal schwannoma at our institution between January 1997 and 2010. The data were gathered retrospectively from medical records and include clinical presentation, tumor location, and post-operative complications. All cases were surgically excised, and they were confirmed to be schwannomas by pathologists with histopathological sections in paraffin stained with hematoxylin–eosin. All cases with a known diagnosis of NF1 or NF2 were excluded. Tumors were classified into 4 groups similar to those used in previous studies according to their relationship to the dura mater: (1) intramedullary, (2) intradural, (3) extradural, and (4) intradural–extradural (dumbbell shaped). Neurological outcome was assessed using the medical research council muscle test, also commonly used for determining, comparing, and classifying motor dysfunction.

Results

Of the 35 patients with spinal schwannomas, 20 (58%) were males and 15 (42%) were females. The mean age of the patients was 47.2 years (range 13–76 years). More precisely, the mean age in cervical lesions was 58.3 years, in the thoracic lesions was 45.7 years, and in the lumbosacral lesions was 49.9 years. The tumors were predominantly isointense on T1-weighted images and hyperintense on T2-weighted
Table 1 – Demographic data.

<table>
<thead>
<tr>
<th>Location</th>
<th>Symptoms</th>
<th>Surgical approach</th>
<th>Surgical resection</th>
<th>Complication</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Pain Motor</td>
<td>Surgical approach</td>
<td>T Subt</td>
<td></td>
</tr>
<tr>
<td></td>
<td>weakness</td>
<td>Post Ant-lat-Ret</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cervical</td>
<td>4 2</td>
<td>Post</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Cervico-thoracic</td>
<td>2 2</td>
<td>Post</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Thoracic</td>
<td>2 2</td>
<td>Post</td>
<td>2 3</td>
<td>MW:2</td>
</tr>
<tr>
<td>Thoraco-lumbar</td>
<td>4 6</td>
<td>Post</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Lumbar</td>
<td>9 4</td>
<td>Post 2</td>
<td>10 2</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>21 14</td>
<td>33 2</td>
<td>30 5</td>
<td>2</td>
</tr>
</tbody>
</table>

MW: motor weakness; T: total; Subt: subtotal; Ant-lat-ret: anterolateral retroperitoneal.

Table 2 – Location and level of 35 spinal schwannomas.

<table>
<thead>
<tr>
<th>Tumor location</th>
<th>Intradural–intramedullary</th>
<th>Intradural–extramedullary</th>
<th>Extradural</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cervical</td>
<td>6</td>
<td></td>
<td></td>
<td>6</td>
</tr>
<tr>
<td>Cervico-thoracic</td>
<td>2</td>
<td></td>
<td></td>
<td>2</td>
</tr>
<tr>
<td>Thoracic</td>
<td>1</td>
<td>3</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Thoracolumbar</td>
<td>1</td>
<td>9</td>
<td>10</td>
<td>13</td>
</tr>
<tr>
<td>Lumbar</td>
<td>10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>2 (6%)</td>
<td>30 (85%)</td>
<td>3 (9%)</td>
<td>35</td>
</tr>
</tbody>
</table>

sequences. The lesions in general had smooth contours, a homogenous appearance, enhanced uniformly, and were well delineated after contrast administration. The tumors in six cases were located in cervical area (17%), four in the thoracic area (11%), two in the cervico-thoracic area (5%), 10 in the thoraco-lumbar area (28%), and 13 in the lumbar area (37%) (Table 1). Two tumors were malign schwannoma; all the others were benign, and there was no

Fig. 1 – (A) Coronal T2-weighted MR image shows the lesion as an increased signal intensity originating from right 5. Lumbar root. (B) Axial T1-weighted MR image shows a hypointense mass at the right 5. Neural foramina. Note the typical dumb-bell shape appearance. (C) Gadolinium-enhanced fat-suppressed axial T1-weighted MR image shows heterogeneous enhancement within the mass.
neurofibromatosis link. In terms of duration, the symptoms ranged from 1 to 60 months (mean 11.4 months). The most commonly observed initial symptom was radicular pain followed by weakness in motor activities. Pain was observed in 60% of the patients, and back pain was observed in 40%. The average follow-up period for all cases was 17.8 months. Of the 35 cases, the schwannomas were intradural–extramedullary in 30 cases (86%), intradural–intramedullary in 2 cases (6%), and extradural in 3 cases (9%) (Table 2). Thirty-three cases were extirpated surgically via partial or total laminectomy through a posterior approach, and every effort was made to prevent postoperative instability. Because of the localization of two tumors which were located in lumbar region (Fig. 1) we preferred an anterolateral–retroperitoneal surgical approach. Total removal was achieved in 30 cases (85%) without recurrence but subtotal removal to avoid nerve damage and bleeding was done in 5 cases (15%). We never observed any vertebral instability or any particular spinal deformity related to the surgical procedure. There were postoperative complications in 6 patients; CSF leak in 1 patient, wound infection in 2 patients, urinary incontinence in 1 patient and motor weakness in 2 patients. Postoperative histological findings confirmed schwannoma in all cases. At the time of discharge, most of the patients seemed to be remarkably improved compared to their preoperative neurological status. Histopathological examination of the tumors in two cases revealed that they were malignant schwannomas in which recurrences were observed after two years and reoperated. Postoperatively they were sent for radiotherapy. In one year follow-up no recurrences were observed. In these two recurrent cases symptoms were similar. Localization of these cases was in thoracolumbar and the lumbar region (Fig. 2).

Discussion

Spinal schwannomas are generally categorized as intradural, extradural, intradural–extradural (dumbbell-shaped), and intramedullary tumors that may occur at any level of the spinal column. In many surgical series, researchers have grouped spinal neurofibromas and schwannomas together under titles like “neurinomas”, “neuromas” or “NSTs”, but it has been suggested by evidence that neurofibromas and schwannomas show different demographic, histological, and biological features that calls for separate classification and further consideration. Both neurofibromas and schwannomas are principally composed of neoplastic Schwann cells, which normally do not exist in the parenchyma of the central nervous system. It has been proposed that the origin of these tumors is the schwann cells that exist in the perivascular nerve plexuses that penetrate the parenchyma of the central nervous system around the pial vessels. According to other theories these tumors may originate from neural crest cells that inappropriately migrate into the parenchyma of the central nervous system during embryonic development, from dorsal or ventral nerve roots with an intraspinal extension, or from pial cells that undergo metaplasia into schwann cells. However, the two tumors present anatomo-pathological differences. Neurofibromas are areas of increased thickness of the nerve, often dumbbell-shaped and situated next to the intervertebral foramina. Microscopically, they present loosely textured areas without Antoni type A patterns, unlike schwannomas, which are characterized by high cellularity, relative lack of Antoni B patterns, and the possibility of mitotic activity with cytologic atypia. Schwannomas are well demarcated, encapsulated, typically round, and attached to the nerve roots. That is why they can be completely resected with minor deficits. Only neurofibromas may undergo malignant change. The “multiple” form of neurofibromas is known as von Recklinghausen’s disease. We have encountered a capsulated, sometimes cystic, regularly shaped, smooth, occasionally bumped tumor. In a few cases,

Fig. 2 – (A) and (B) The lesion was totally resected and there is no signal abnormality corresponding to a residual lesion on coronal T1 W (A) and contrast enhanced axial T1W images.
it originated from the motor anterior root assuming a ventral or ventro-lateral topography with respect to the medulla, while more frequently it originated from the sensory root assuming a posterior or postero-lateral topography. In the literature, patients with type 2 Neurofibromatosis (NF2) seem to develop malignant forms in 30–50% of the cases. In the malignant forms the manifesting symptoms are often more defined and worsen rapidly. NF2 is characterized by an alteration of the gene located on the chromosome 22q that predisposes the patient to the development of multiple tumors in the central and the peripheral nervous system. Although a major incidence in the cervical and lumbar tracts has been reported, the tumor presents an ubiquitous evolution in the spine. In our study, a higher incidence was observed in the lumbo-sacral spine (37%). In the literature, it is reported that 70–80% of spinal schwannomas are intradural in location, and others, which account for another 15%, extend through the dural aperture as a dumbbell mass with both intradural and extradural components. Intramedullary schwannomas are extremely rarely observed. Accordingly, we saw two cases that were intradural-intramedullary schwannomas in our series. In the initial stages, the root pain is attributed to the disturbance of nerve conductivity due to the direct or indirect irritation of nerve root or root compression by the tumor. However, motor weakness almost never occurs as an initial symptom in the lumbosacral region. Motor weakness of the lower extremity may not be apparent until the later stage, as in the case of patients with lumbar canal stenosis. Although complete resection of spinal nerve sheath tumors may be feasible, some cases have been resected incompletely. Total resection can be hindered in two ways: one is attachment to the spinal cord due to hemorrhage, inflammation, or subpial localization; the other is adherence of critical structures to extradural components such as the vertebral artery outside the spinal canal in the cervical region. In our study, total resection was not achieved in either case because of these obstacles. However, a better understanding of the anatomy of surrounding structures and meticulous surgical techniques could eliminate these hindrances. In case of a residual tumor, observation for a long time is necessary. The level of angulations in the course of the development of kyphosis following laminectomy is often correlated with removal of facet joints. Even though facets are not removed at all, a rounded kyphosis might develop gradually. With bilateral removal of facets at one or more levels, acute angular kyphosis is likely to occur. Unilateral facetectomy can predispose to angular kyphosis, and scoliosis with the angular kyphosis may cause spinal cord compression although recurrent tumor is absent. Osteoplastic laminoplasty for spinal tumor is a few children was reported by Abbott et al.; however, there were not enough data to establish the efficiency of this procedure. Another study by Kim et al. showed that the laminoplasty prevents postoperative spinal deformities after removal of spinal cord tumors in 16 cases of laminoplasty compared with 89 cases of laminectomy. Kawahara et al. performed recapping T-Saw laminoplasty for spinal cord tumors in 24 patients. They reported that complications such as postoperative spinal canal stenosis, facet arthrosis, or kyphosis were not observed. The result of schwannoma treatment is related to preoperative neurological condition of the patient. The symptoms of our patients, with regards to pain and motor weakness, improved by 95%. Complete clearance of neurofibromas and schwannomas that are not linked to neurofibromatosis is generally curative. However, tumors with extensive paraspinal involvement subtotally resected have a certain propensity to recur. Deficits resulting from sacrifice of the involved nerve roots are often minor and may well be tolerated. In our study, there were two recurrences in 5 cases treated by subtotal removal to avoid the nerve root injury.

**Conclusion**

Spinal schwannomas may occur at any level of the spinal axis and are typically intradural. The most commonly cited clinical presentation is pain. Even though they are classified as spinal nerve sheath tumors with neurofibromas, schwannomas are different from neurofibromas regarding clinical outcome and histopathology. As we experienced, when compared with schwannomas in neurofibromatosis cases, most spinal schwannomas in non-NF cases can be resected completely without inducing deficits or with only minor postoperative deficits. Radiotherapy is effective in histopathologically confirmed malignant schwannomas as we observed in our two recurrent cases.

**Conflict of interest**

The authors declare no conflict of interest.

**References**